382 Quantifying publication bias in cystic fibrosis clinical trials

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Background: The practice of evidence-based medicine depends upon the publication of high quality clinical studies. However, publication bias may act to exclude some trials from contributing to the knowledge base. The influence of such biases upon the evidence base is often unknown as clinicians have no way of knowing the results of unreported studies. ClinicalTrials.gov was commissioned in 2000. In 2005 the ICMJE announced that publication of trials would be limited to trials registered with a public registry prior to commencing.
Aim: We aimed to quantify reporting biases present within the CF literature.
Method: We interrogated ClinicalTrials.gov using 'cystic fibrosis' as the keyword, limited to interventional studies that had been closed. We searched PubMed, the Cochrane Central Register of Controlled Trials and Google Scholar for publications using ClinicalTrials.gov ID, study title and Chief Investigator name. We documented publication date and study sponsor. A survival analysis was performed using R (version 2.12.1).
Results: In total 142 studies were identified (completed 1998–2010) of which 59 were published. Median time to publication was 2.09 years.

Proportion published by end of year

Year	Proportion of studies published at the end of each year following completion, after taking into account right censored studies
1	6.7%
2	24.5%
3	47.9%
4	56.6%
5	59.0%

We performed a similar survival analysis with categories of sponsor as subgroups.
Conclusion: The publication of results of clinical trials, which contribute to the evidence base on which clinicians depend, are often delayed.

383 Upper airway pathology in children with cystic fibrosis (CF)

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Upper airway (UAW) pathology in CF has been neglected and can be a source of bacterial infections of lower airway, and significantly decrease the quality of patients life.
Objectives: To study the frequency of chronic rhinosinusitis (CRS) in children with CF, correlation of genotype and ENT disorders and effectiveness of dornase alfa [inhaled via a special nebulizer (PARI SINUS™)].
Methods: The study included 103 patients with CF (48 boys and 55 girls) aged from 1 to 17 years. 14 children received a 1-year dornase alfa, twice a day at a dose of 2.5 mg. Special functional diagnostic of ENT were nasal endoscopy and CT scan.
Conclusion: CRS was diagnosed in 83 patients (82% of total), in which 56 patients (68%) – with nasal polyps. Predominant clinical signs were chronic nasal congestion, recurrent rhinitis in 83 (100%), rhinorrhoea in 53 (64%), mouth breathing, anosmia in 19 (23%), facial pain in 15 children (18%). Severe disease of Shwachman score was observed in 36 out of 103 patients (35%), all of them were with nasal polyps and delF508 mutations. All children without CRS (20 patients) belonged to the younger age group (up to 3 years). The treatment of dornase alfa revealed significant improvement in nasal breathing and reduction of polyps of investigated group.

384 Pregnancy and motherhood in women with cystic fibrosis: experience and outcomes in a regional adult UK centre

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As the median survival for CF continues to improve more women face decisions regarding pregnancy. The aim of this study was to review the experience of pregnancy and assess both maternal and fetal outcomes.
A retrospective review of the case notes of women who became pregnant between 1990 and 2009 was performed. Maternal lung function, weight, number of clinic visits and days of intravenous antibiotics were recorded for 4 years pre pregnancy, during pregnancy and 4 years post pregnancy.
35 women (median age 24 yrs, FEV_1 62% and BMI 20.9) became pregnant and delivered 50 children. 13 had pre-existing CFRD and 6 women developed gestational diabetes. Median (range) maternal weight gain during pregnancy was 6.5 kg (−2.9 to 15.3 kg). There was a significant increase in number of clinic visits ($p = 0.009$) and requirement for iv antibiotics during pregnancy and up to 2 yrs post delivery ($p = 0.013$). Women with CFRD had lower weight gain during pregnancy (5.3 vs. 6.8 kg, $p = 0.024$), lower birth weight babies (2.1 vs. 2.96 kg, $p = 0.002$) and shorter gestation (34 vs. 37 weeks, $p = 0.001$). A prepregnancy FEV_1 less than 60% predicted was associated with lower maternal weight during pregnancy (49.9 vs. 59.4 kg, $p = 0.04$), shorter gestation (34 vs. 36 weeks, $p = 0.018$), lower birth weight (2.35 vs. 2.67 kg, $p = 0.022$) and a significant decline in FEV_1 post pregnancy ($p = 0.03$). Seven women died post pregnancy.
The course of pregnancy cannot be predicted in any individual and all women should anticipate more intensive antepartum and postpartum treatment. The presence of pre-existing CFRD and a prepregnancy FEV_1 of <60% predicted are associated with significantly worse outcomes.

385 Smoking, alcohol and drug use in cystic fibrosis patients

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Introduction: As young adults living with a chronic respiratory illness, cystic fibrosis (CF) patients are more prone to the harmful effects of tobacco smoke, excessive alcohol and recreational drugs. We wished to look at the prevalence of smoking, drugs and alcohol use in our cohort of adult CF patients.
Methods: Using a structured questionnaire, we surveyed 98 consecutive CF patients (mean age 28 yrs [range 18–66], 49 female) attending our large adult CF unit looking at smoking, alcohol, and recreational drug use.
Results: Although 80% had never smoked tobacco, 8% were ex-regular smokers (average 7.8 pack-yrs [range 2–16]), 4% were regular smokers, 5% were occasional (not daily) smokers, and 3% smoked rarely. Only 2% of the smokers had used some form of aid to try to quit. 86% took alcohol, with 66% using it less than once a week but 33% more frequently (average 15 units per week [range 1–40]). Only 1 patient had attended a formal rehabilitation programme for excessive drinking. Finally, with respect to recreational drug use, 13% had ever used drugs and 3% took drugs at least once a week (the commonest being cannabis, followed by LSD and cocaine). None had attended any rehabilitation programme.
Conclusion: Smoking, excessive alcohol and recreational drug use is a real problem among the adult CF population and it is important that these issues are discussed and explored at periodic reviews within the CF team to identify patients who are most vulnerable. The long-term deleterious impact of unhealthy life choices should be reiterated at every opportunity to high-risk patients, and adequate facilities should be made available to those who want help to deal with these habits.